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Surgical treatment of Löffler's eosinophilic endocarditis

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SUMMARY Löffler's eosinophilic endocarditis is a rare cardiac disease which causes endocardial damage and mural thrombosis, leading to restriction of the ventricular cavity and atrioventricular valvar regurgitation.

A case of a middle-aged woman with Löffler's endocarditis is presented. After the removal of the restrictive thrombotic tissue from the left ventricular cavity and replacement of the mitral valve the patient was asymptomatic, but died 14 months after operation because of thrombosis of the prosthesis. Nevertheless, surgical treatment of the mechanical pump disturbance is recommended in addition to drug treatment.

Löffler's eosinophilic endocarditis is a rare cardiac disease[1–6] with a poor prognosis.[1 7–10] Endomyocardial damage and fibrosis as well as mural thrombosis restrict the filling and output of the ventricles and also interfere with the function of the atrioventricular valves, leading to significant valvar regurgitation.[1 2 4 6–8 11] Conservative treatment with cortisone, immunosuppressive, or cytotoxic agents is often disappointing or causes remissions of short duration.[1 3 4 10] Hence the surgical correction of the life-threatening mechanical disturbance of cardiac function seems worth while.[12]

We present a case of successful surgical treatment of left ventricular mural thrombosis and mitral regurgitation caused by this disease. The patient, however, died 14 months after operation because of thrombosis of the mitral prosthesis.

Case report

A 45-year-old woman who had had bronchial asthma for three years suffered a massive allergic reaction with idiopathic angioneurotic oedema. Her white cell count was $15{\cdot}1 \times 10^9/l$ with an eosinophilia of 62 per cent. Oral cortisone was given, but three months later her dyspnoea became worse, appearing both at exercise and rest. In a district hospital, lung râles and a third heart sound were found on auscultation. Peripheral cyanosis, leg oedema, and hepatomegaly were also found. There was pulmonary venous congestion and fluid in both pleural spaces on the chest *x*-ray film. The treatment of cardiac failure was begun with idgoxin and diuretics; steroids were continued as 20 mg prednisolone daily.

This regimen was maintained for two months but the dyspnoea became worse and the patient was sent to Oulu University Central Hospital. She was now very breathless at rest. Her lips were cyanotic and peripheral pitting oedema and hepatomegaly were found. The neck veins were distended and both a and v waves were visible. The carotid pulse volume was low and a left parasternal lift was palpable. On auscultation an accentuated pulmonary component of the second heart sound, a third heart sound, and a loud apical systolic murmur of mitral valve regurgitation were heard.

The electrocardiogram showed left axis deviation, a QS pattern in V1-3, and P mitrale. There was slight right-sided cardiac dilatation, pulmonary venous congestion, and an obvious right pleural effusion on the chest *x*-ray film (Fig. 1a).

Echocardiography showed that the anterior mitral leaflet was normal, but the posterior leaflet was not found. The left atrial diameter was slightly increased. The left ventricular dimensions and function were within the normal range, but the left ventricular posterior wall was thicker than normal, up to 22 mm. The interventricular septum contracted vigorously with a wide amplitude, suggesting mitral valve regurgitation.

The patient was afebrile and her erythrocyte sedimentation rate was 29 mm/h. Haemoglobin was 139 g/l and the leucocyte count $12{\cdot}9 \times 10^9/l$. No eosinophils were found in the differential or in the absolute count. The serum electrolyte and enzyme concentrations were within normal ranges except for serum lactic dehydrogenase and alkaline phosphatase which were slightly raised.

The data from cardiac catheterisation and quan-

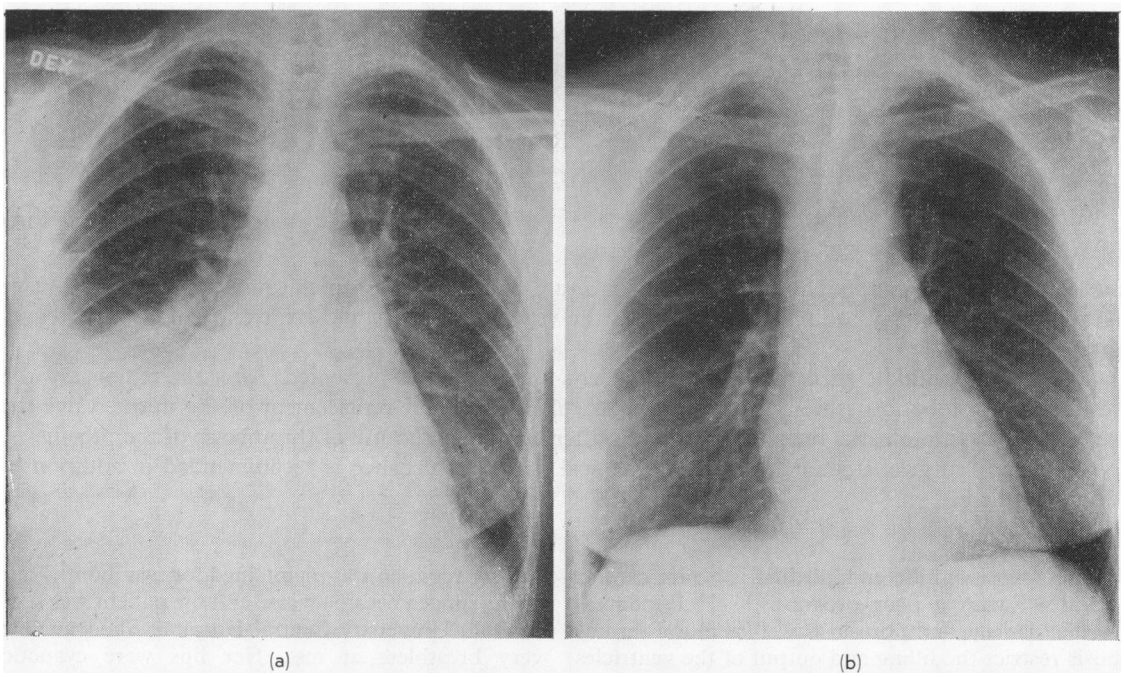

Fig. 1 *The anteroposterior chest x-ray film of the patient before operation (a). The heart size is normal, but the pulmonary veins are congested and a large right-sided pleural effusion is visible. After operation the chest x-ray film returned to normal (b).*

titative left ventriculography are presented in the Table. In selective biplane left ventricular cineangiography the left ventricle was slightly hour-glass shaped, with an irregular inner wall contour (Fig. 2a). The left ventricular end-diastolic volume was smaller than normal, but the ejection fraction was within normal limits and left ventricular contraction was symmetrical. Grade 4/4 mitral valve regurgitation was found and the left atrium was slightly dilated.

At the operation the anterior mitral valve leaflet was found to be normal, but the chordae tendineae near the right commissure were shortened, hindering the closure of the valve. The inner wall of the left ventricular cavity was covered throughout with friable light grey thrombotic tissue, which totally encapsulated the posterior mitral valve leaflet with its chordae and papillary muscles. The thickness of the abnormal tissue was up to 1 centimetre. Hence the left ventricular cavity was smaller than normal and hour-glass shaped.

The abnormal thrombic masses were removed manually so that smooth and pale endocardium became visible. The mitral valve was excised and replaced with a Björk-Shiley tilting disc valve prosthesis.

Table *Data from cardiac catheterisation and quantitative left ventriculography*

	Before operation	*One year after operation*
Right atrial pressure (mmHg)		
Mean	18	7
a wave	23	11
v wave	21	8
Right ventricular pressure (mmHg)		
Systolic	61	34
End-diastolic	20	8
Pulmonary artery pressure (mmHg)		
Mean	45	20
Systolic	60	30
Diastolic	39	14
Pulmonary wedge pressure (mmHg)		
Mean	29	14
v wave	38	18
Left ventricular		
Systolic pressure (mmHg)	92	111
End-diastolic pressure (mmHg)	26	14
End-diastolic volume (ml)	69	83
End-diastolic volume index (ml/m²)	42	50
Stroke volume (ml)	47	44
Forward stroke volume (ml)	28	44
Ejection fraction (%)	68	53
Mitral valve regurgitation		
Absolute (ml/beat)	19	0
Regurgitation fraction (%)	40	0
Cardiac index (l/min per m²)	1·9	2·5

The abnormal mass weighed 20 g and microscopically consisted of thrombotic tissue. Bacterial and fungal cultures from it were negative. The histology of the atrial appendage and the mitral valve were normal, but no specimen was taken from the left ventricular endocardium.

The patient recovered normally and was well one year after operation. Dyspnoea occurred only during unusual exercise and she had resumed a normal life. The signs of right ventricular failure had disappeared and on auscultation normal prosthetic sounds without murmurs were heard. Her leucocyte count was $8{\cdot}3 \times 10^9/1$, of which 1 per cent were eosinophils. The chest *x*-ray film (Fig. 1b) and the electrocardiogram were normal. On the echocardiogram the left ventricular posterior wall thickness was normal, as were left ventricular dimensions and function. The left atrial diameter had also decreased slightly. Cardiac catheterisation now showed pressure measurements that were nearly normal (Table) and cineangiography disclosed a normally shaped left ventricular cavity with a competent mitral valve prosthesis (Fig. 2b). She was still taking digoxin, diuretics, anticoagulants, and prednisolone, 10 mg daily.

Fourteen months after operation she became acutely dyspnoeic with pulmonary oedema and died within several hours in a district hospital. Necropsy showed thrombosis of the mitral prosthesis as the cause of death. Macroscopically, the left ventricular endocardium seemed unaltered compared with findings at operation. Microscopically, the endocardium was 2 mm thick, consisting of collagen and hyaline material but with no eosinophilic infiltrate. Similar changes were found patchily in the right ventricle. These changes are typical of the chronic fibrous stage of Löffler's endocarditis. Mural thrombosis was not found in the ventricles or atria.

Discussion

The thrombotic involvement of the left ventricular inner wall encompassed the posterior mitral valve leaflet so that the dominant feature was severe mitral valve regurgitation, together with left ventricular cavity restriction. Forward cardiac output was therefore very low and symptoms were severe. Eosinophils had totally disappeared, perhaps at least partly as a result of cortisone therapy, and so the problem was principally the mechanical disturbance in cardiac function and not the active systemic disease. On the basis of the catheterisation data, the right ventricular failure was considered to be secondary to mitral regurgitation, and indeed the symptoms and signs of right ventricular failure completely disappeared after operation. Löffler's endocarditis may affect the left ventricle alone,[2 11] according to Brockington and Olsen in 39 per cent of cases. Hence in our patient left ventricular thrombectomy and mitral valve replacement were deemed sufficient to restore cardiac function.

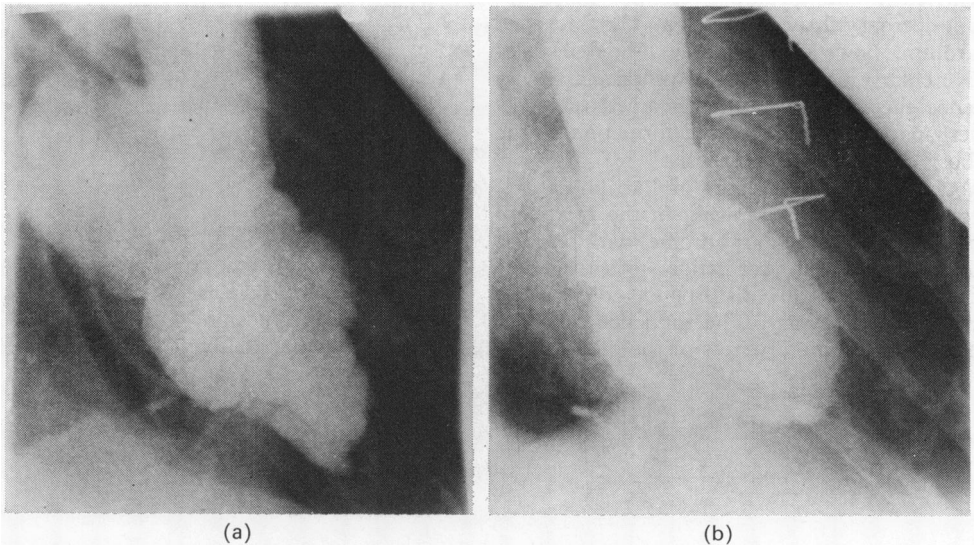

Fig. 2 *The preoperative end-diastolic left ventriculogram in right anterior oblique position (a). The inner contour of the left ventricular wall is irregular, suggesting intracavitary thrombosis. Severe mitral regurgitation opacifies the left atrium. After operation the left ventricle is normal (b).*

Because the macroscopical changes of the left ventricular endocardium were slight it was not removed. It would not have been easy to do and a raw muscle surface would probably have been more susceptible to the formation of thrombosis than smooth endocardium.

The surgeon inserted a Björk-Shiley valve prosthesis, because this type of valve prosthesis was in use in our hospital at that time. Being less prone to thrombosis a bioprosthetic valve might have been preferable, additionally because mural thrombosis of the left ventricle is more likely to hinder the opening of the Björk-Shiley tilting disc than the opening of a bioprosthetic valve. Antiplatelet drugs might have reduced the risk of the thrombosis of the valve prosthesis, but would have had less effect on the risk of formation of new left ventricular mural thrombosis.

To our knowledge there has been no previous report of the surgical treatment of Löffler's endocarditis at the stage of thrombosis. Dubost *et al.*[12] reported five successful cases, but all the patients had obvious fibrosis of the endocardium extending to the mitral valve, a later stage of the disease than that seen in the present case and one in which excision of the endocardium is mandatory if good mechanical function is to be restored. In Davies's endomyocardial fibrosis, which is obviously a later stage and a more chronic form of Löffler's endocarditis,[2 6 7 9 11] scar excision with atrioventricular valve replacement has been performed.[13]

In our patient, the macroscopical changes in the left ventricular endocardium were not more apparent at necropsy than at operation. The thickened endocardium, however, obviously hindered postoperative enlargement of the left ventricular cavity and hence the cardiac output remained relatively low, perhaps contributing to the formation of the thrombosis of the valve prosthesis.

Although in a recent report of Parrillo *et al.*[10] hydroxyurea appeared promising in the treatment of the hypereosinophilic syndrome with cardiac involvement, surgical correction of the life-threatening mechanical disturbance of cardiac function seems necessary. The influence of endocardial excision on the progress of the disease is as yet obscure.

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